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Sinus Histiocytosis with Massive Lymphadenopathy (Rosai-Dorfman Disease): A Case Report with Review of Literature

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ABSTRACT

Sinus Histiocytosis with Massive lymphadenopathy, also commonly known as Rosai-Dorfman Disease is a rare non neoplastic histiocytic disorder first described by Rosai and Dorfman, seen predominantly in childhood and young adults. It is characterized by fever, painless massive cervical lymphadenopathy and elevated ESR levels. RDD is often misdiagnosed as lymphoma, tuberculosis or metastatic malignancy, so it is important to distinguish it from these conditions as the modes of treatment are different. Here we report a case of 18 years old male presenting with massive lymphadenopathy. Morphological findings demonstrated Emperipolesis consistent with a diagnosis of Rosai-Dorfman disease. Clinical and cytological aspects of the disease are discussed as a rare cause of lymphadenopathy.

Keywords: *Sinus Histiocytosis with Massive Lymphadenopathy (SHML), Rosai-Dorfman Disease (RDD), Emperipolesis, Lymphadenopathy.*

INTRODUCTION

Sinus Histiocytosis with Massive lymphadenopathy also known as Rosai-Dorfman Disease is a rare benign histioproliferative disorder [1] first identified and characterized by Rosai and Dorfman as a separate entity in 1969 [2]. It is a disorder of unknown etiology occuring in first and second decades of life characterized clinically by painless bilateral cervical lymph node enlargement associated with fever and elevated ESR levels with characteristic microscopic findings [3]. The disease has a wide geographical distribution with most of the cases reported from US and Western Europe along with African continent [4] with few number of cases reported from this part of world. The disease is usually a self limiting one, however depending upon the severity of disease, different treatment modalities ranging from medical to surgical excision may be used [5]. Here we report a case of 18 years old male who presented with massive bilateral cervical lymphadenopathy.

CASE REPORT

An 18 years old male presented in our hospital with history of low grade fever associated with bilateral, non tender, multiple enlarged cervical lymph nodes of 4 months duration. There was no associated history of pain, weight loss, night sweats or respiratory tract infections. He had no significant past history of any major illness or family history of tuberculosis or blood disorders. There was no associated organomegaly and rest of the systemic examination was unremarkable.

Clinical examination showed multiple, bilateral enlarged cervical lymph nodes which were discrete and non tender and ranging in size from 1.5 to 6.0 cms in diameter. Laboratory investigations revealed hemoglobin 10.2 gm/dl, total leucocyte count 15,200/cu mm and ESR levels 82mm/1st hour (Westergrens method). Serological tests for HbsAg and HCV, Sputum for AFB and tuberculin test were negative. Liver function tests and renal function tests were within normal range. Chest X ray, Ultrasonography and CT scan chest and abdomen were normal. Patient was referred to Pathology section for Fine Needle Aspiration Cytology (FNAC) of cervical lymph nodes.

FNAC of bilateral cervical lymph nodes was performed using 22 gauge needle and 10 ml plastic syringe with a detachable syringe holder (Franzen Handle). Three alcohol fixed smears were prepared, first smear was stained with Papanicolaou stain, second with Giemsa stain and third one was kept unstained for any further required stain. Microscopic examination revealed cellular smears comprising of numerous histiocytes with abundant pale cytoplasm and single to multilobed nuclei without nuclear atypia or grooving (Fig 1). The cytoplasm contained variable number of lymphocytes (Emperipolesis) and neutrophils in some areas (Fig 2,3). The background was partly haemorrhagic containing lymphocytes, plasma cells, neutrophils and few binucleated macrophages. Based on these typical findings, a diagnosis of Rosai-Dorfman Disease was made. Subsequent excision biopsy and histopathological examination of the lymph node confirmed the cytological findings and a final diagnosis of Sinus Histiocytosis with Massive lymphadenopathy (Rosai-Dorfman Disease) was made.

Patient was counselled by the treating physician about the nature of disease and was put on oral corticosteroids. In the follow up, response to therapy was excellent and the lymph nodes gradually decreased in size after a period of 4 weeks. No side effects were noted. The patient is still on follow up.

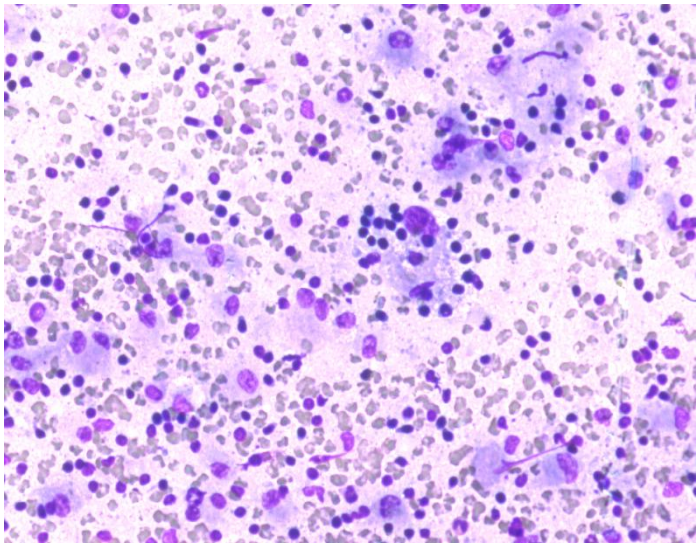

Fig 1: Microphotograph showing numerous mono and binucleated histiocytes showing lymphophagocytosis (40x, Giemsa)

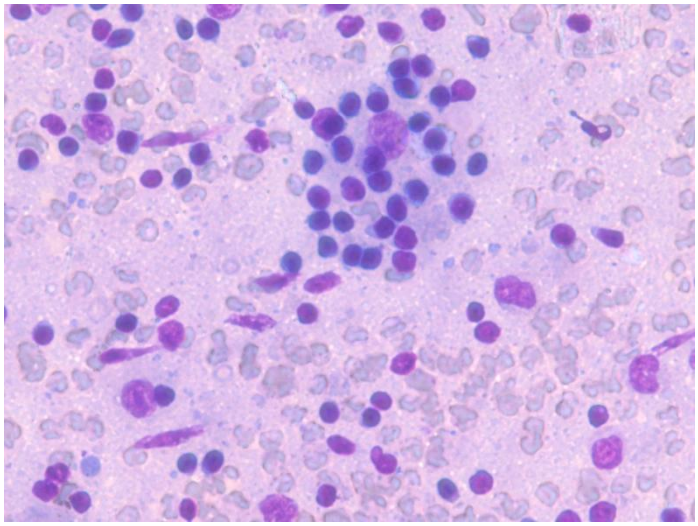

Fig 2: Microphotograph depicting emperipolesis with numerous intact lymphocytes ingested by histiocyte (80x,Giemsa)

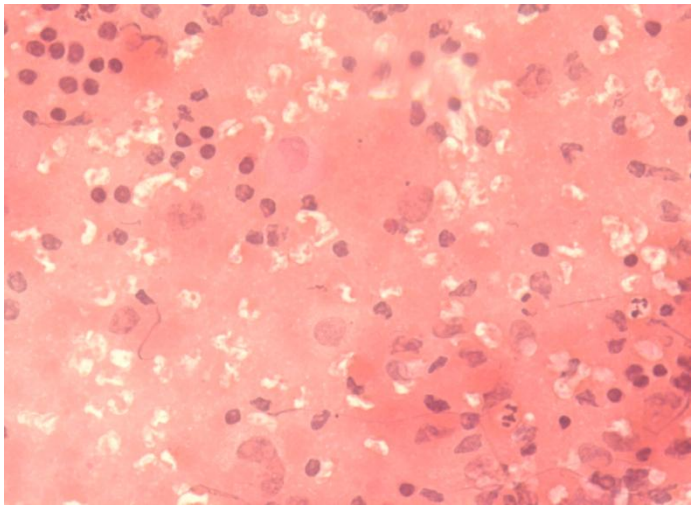

Fig 3: Microphograph depicting emperipolesis (80x,PAP)

DISCUSSION

SHML or RD Disease is a rare benign, self limiting histiocytic disorder occurring mostly in children and young adults with a tendency for self remission Cardinal features include painless cervical lymphadenopathy, fever and elevated ESR. It mostly occurs in whites, more commonly in first and second decades of life and more in females. [6]. More than 90% of patients with RD disease present with massive bilateral, mobile and non tender cervical lymphadenopathy. Axillary, inguinal and mediastinal lymph nodes can be involved in 30-40% cases. Extranodal involvement may be seen in upper aero digestive tract, salivary glands, orbit, skin and paranasal sinuses [7]. Sodhi et al [8] reported a case of RD disease with unusual presentation of diffuse and massive retroperitoneal lymphadenopathy.

Etiology of the disease is still unclear and several theories have been proposed as the cause of the disease including Epstein Barr virus and Human Herpes Virus (HHV-6) or immunological factors due to presence of disorders of humoral immunity and decreased cell mediated immunity [9]. Emperipolesis or lymphophagocytosis is one of the main diagnostic features characterized by the presence of intact lymphocytes engulfed within the cytoplasm of histiocytes. The internalized lymphocytes are usually located within the cytoplasmic vacuoles. Ultrastructurally, histiocytes lack Birbeck granules and viral particles. Besides cytomorphology, the histiocytes on immunostaining show positivity for S100 protein, CD14, CD33 and CD 68 in cytological smears [10].

Pathological differential diagnosis of Rosai-Dorfman Disease includes reactive sinus hyperplasia (no emperipolesis, S100 negative), Langerhans cell histiocytosis (positive for both S100 and CD1a), Hodgkins disease, metastatic carcinoma, malignant melanoma and lymphoma. Diagnosis of RD disease requires correlation of clinical findings, laboratory findings and cytomorphological findigs along with immunostaining [11].

Treatment has not yet been defined. However the prognosis is excellent in most cases. Complete spontaneous regression is known to occur. Only few cases of progression to malignant melanoma or sarcoidosis have been documented yet [4]. Complete remission frequently occurs over several months or years, either spontaneously or with administration of steroid treatment, as in our case. Corticosteroid therapy allows for transient improvement of the disease. Radiotherapy is reserved for active life threatening forms of disease refractory to corticosteroids and surgery [7].

CONCLUSION

Sinus histiocytosis with massive lymphadenopathy or Rosai-Dorfman disease should be considered in the differential diagnosis of patients especially children and young adults presenting with painless lymphadenopathy in head and neck region. Clinicians in coordination with pathologists should be able to recognize the entity keeping in consideration the characteristic clinical and microscopic findings, since if properly diagnosed and managed, unnecessary diagnostic modalities and mismanagement can be avoided.

CONFLICT OF INTEREST: Nil

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